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ΑΡΧΕΙΑ ΕΛΛΗΝΙΚΗΣ ΙΑΤΡΙΚΗΣ 2016, 33(2):280–281

Medical Imaging Quiz – Case 39

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A 40-year-old woman presented to the emergency department due to high fever, cough and severe fatigue for 5 days. Two weeks before admission, she had travelled to India. She had a history of type 1 diabetes. In admission, she complained of acute dyspnea and thoracic pain. Physical examinations showed temperature of 38 °C, pulse rate: 96/min, respiratory rate: 16/min, blood pressure: 110/70 mmHg, and pharyngeal erythema without exudates. She underwent a full laboratory investigation, including full blood count, biochemical tests and chest x-ray that revealed consolidation in the lower left lobe. Chest computed tomography (CT) showed an oval-shaped solid mass with well-defined borders in the lower left lobe. The mass had homogeneous density with minimal contrast enhancement (fig. 1). The rest of the lungs were clear, and no mediastinal lymphadenopathy was seen. According to patient's history and clinical appearance, the differential diagnosis consisted of infection (tuberculous abscess), benign or malignant lesion. Fine core biopsy confirmed the diagnosis.

Comment

Neurofibromas are benign peripheral nerve sheath tumors, usually solitary and sporadic; however there is an association with neurofibromatosis type 1 (NF1). Three types have been described: localized neurofibroma, diffuse neurofibroma, plexiform neurofibroma (pathognomonic of NF1). Localised neurofibromas consist the most common type of neurofibroma (approximately 90%). The majority are solitary and sporadic, not associated with NF1. Neurofibromas are benign, slow growing neoplasms, usually <5 cm in size at presentation, composed Schwann cells and fibroblasts, containing a rich network of collagen fibres. Primarily they affect superficial cutaneous nerves; however, occasionally affect larger deep-seated nerves. Neurofibromas might undergo malignant transformation into a malignant peripheral nerve sheath tumour. Peak presentation is between 20 and 30 years of age with no sex predilection.

Clinical presentation: Spinal neurofibromas are often asymptomatic. If symptoms are present, they usually include pain and or radicular sensory changes due to the typical location along the dorsal sensory roots. Weakness is less common. Myelopathy may occur if the lesion is large.

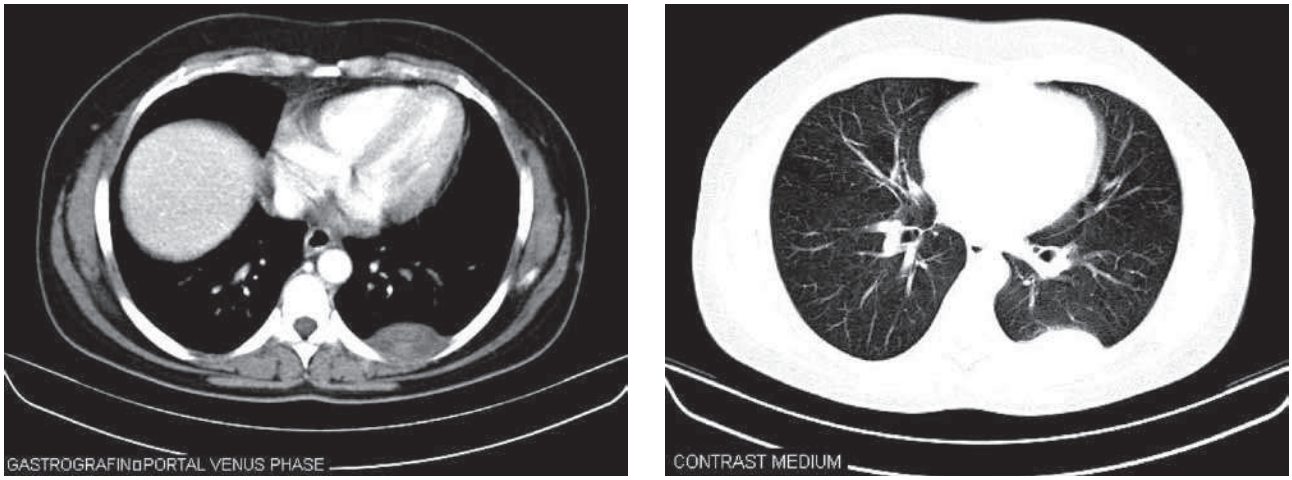

Figure 1. CT reveals oval-shaped solid mass with well-defined borders in the lower left lobe. The mass has homogenous density with minimal contrast enhancement.

Radiographic features: These tumours present as a well-defined hypodense mass with minimal or no contrast enhancement on CT. On magnetic resonance imaging (MRI), they usually are T1 hypointense and T2 hyperintense with heterogeneous contrast enhancement. A hyperintense rim and central area of low signal resulting in a target sign may be seen; this is thought to be due to a dense central area of collagenous stroma. Although this sign is highly suggestive of neurofibroma, it is occasionally also seen in schwannomas and malignant peripheral nerve sheath tumours.

Surgery is the treatment of choice for symptomatic lesions. However, as neurofibromas tend to encase the nerve roots, they usually cannot be dissected from the parent nerve; deep-seated lesions are therefore often managed conservatively. Local recurrence after excision is uncommon and malignant transformation is rare (5–10%), which may be indicated by rapid growth.

The main differential diagnosis for neurofibromas includes spinal schwannoma and spinal meningioma.

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Answer: Neurofibroma